PRENATAL CAUSES OF DEAFNESS

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I. INTRODUCTION

II. MATERNAL RUBELLA

- Affects on the fetus
- Remediation

III. RH FACTOR

- Affects on the fetus
- Remediation

IV. DRUGS

V. OTHER CAUSES

- False labor
- Influenza
- Congenital syphilis
- Diabetes
- Beri-beri
- Glandular fever
- Cretinism
- Malabsorption syndrome
- Toxemia of pregnancy

IV. CONCLUSION

I. INTRODUCTION

When considering the causes of congenital perceptive deafness, it is customary to consider them in a chronological sequence. They fall into three main groups. First is the hereditary group, due to genetic influences. Second is the pre-natal group, due to a variety of noxious influences upon the developing embryo. Third is the perinatal group, due to any one of a number of accidents at the time of birth itself, shortly before birth, or within the earliest hours or days after birth.[1]

In a census taken during the 1920's, 38.6 per cent of 35,026 deaf Americans were deaf because of prenatal causes.[2] This is one third of all those responding to the questionnaire attributing their deafness to a prenatal factor.

Many prenatal causes of deafness have been enumerated by researchers. It is stated that these causes are:

> . . . Maternal infection, especially virus diseases:
> rubella glandular fever
> influenza Asian influenza
> Maternal nutritional deficiencies:
> the malabsorption syndrome
> beri-beri diabetes
> Toxemia of pregnancy
> endocrine—cretinism[3]

Another resource agrees with the above causes and adds a few more. These are: ". . . mumps, the Rh factor, certain drugs taken by the mother during pregnancy, pathological condition of the fetus (such as erythroblastosis fetalis), development anomalies, and maternal syphilis."[4]

It is the purpose of this research paper to discuss the great "variety" of prenatal causes of deafness in more detail.

II. MATERNAL RUBELLA

Rubella (German measles) is harmful to the developing fetus if it is contracted during the first three months of pregnancy. "A woman who contracts German measles during . . . this 'high risk' period, has only a 50 per cent chance of having a normal baby."[5] The pregnant woman who has rubella may develop a rash and be slightly ill. It is often the case that the mother is not aware of having measles. When this happens, the deafness of her child is often thought to be inherited and the actual cause remains unknown.[6]

"Deafness due to rubella has been recognized only since about 1947. . . ."[7]

> . . . The deafness is perceptive and may be unilateral or bilateral, severe or moderate. According to Clayton Jones (1947) and Hopkins (1949) there is no obvious relation between severity of the mother's rubella and the degree of the child's deafness and other defects.[8]

Affects on the Fetus

The stage of embryonic life at which the mother is affected by rubella has a great bearing on the damage that is done to the fetus. The damage is greatest soon after conception.[9]

> Gregg (1941) noted the association between congenital cataract and maternal rubella occurring

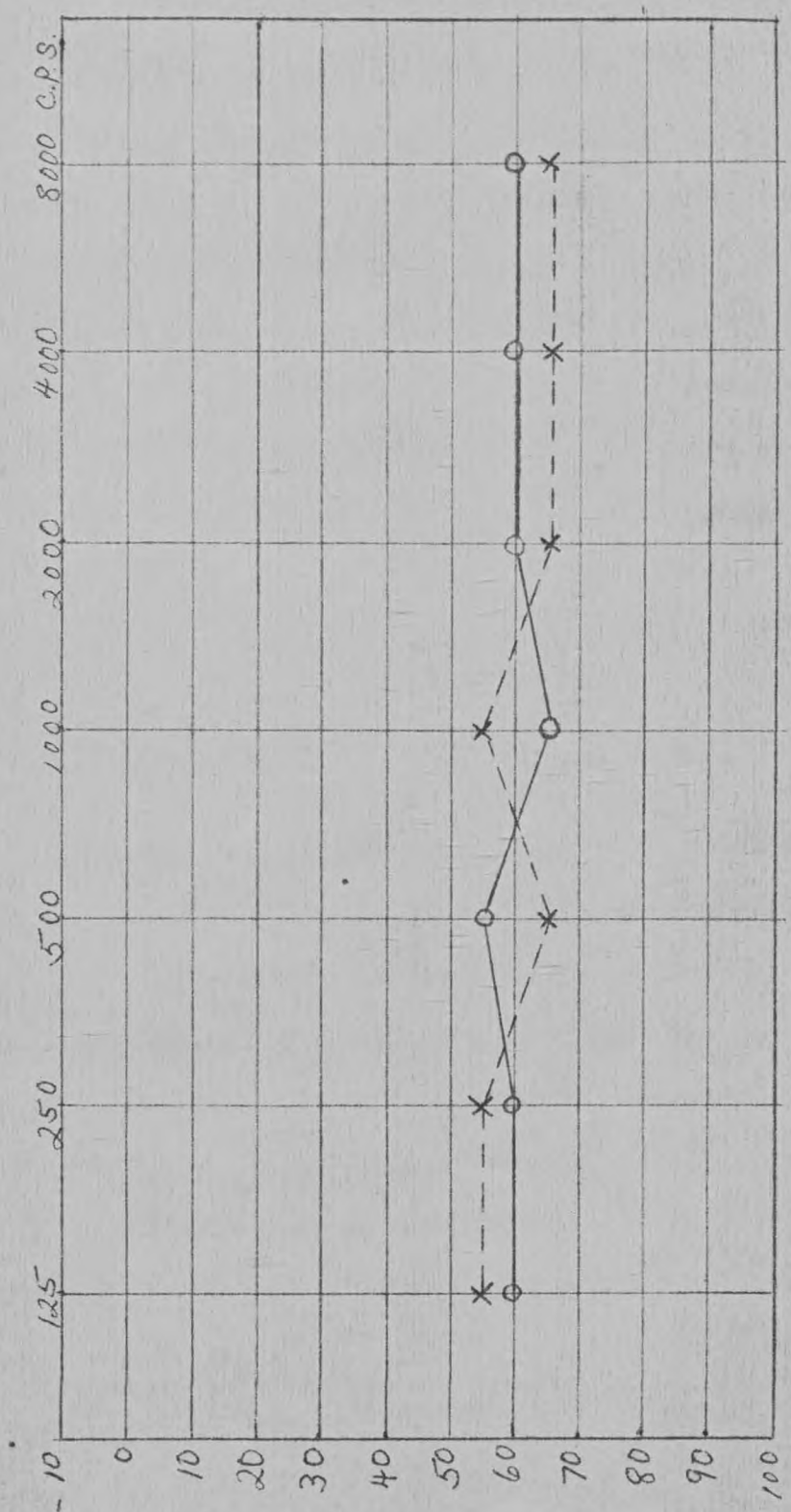
> in the first few months of pregnancy. The association was confirmed by Swan and his associates who also noted the incidence of other defects, mainly perceptive deafness and cardiac defects.[10]

Another researcher categorized the damage done to the fetus with the stage of embryonic life it was in at the time of infection. He concluded that infection occurring in the first six weeks of embryonic life affects the ears, eyes, heart, and maybe other parts including the central nervous system. After the first six weeks, infection will damage the cochlea and may cause some growth retardation.[11]

> . . . It has been said that if it [rubella] developes in the first month of pregnancy, one child in every two is likely to be handicapped—by deafness, blindness, heart disease or mental deficiency. This high incidence is reduced to one-in-four during the second month, one-in-six during the third.[12]

"Since the cochlea [when affected] is usually affected throughout its length, from base to apex, the hearing loss tends to be uniform for all frequencies."[13] The hearing loss is characteristically "flat", and commonly at a level of about 60 decibels (see figure 1 on page 5).[14]

It is also stated that: "The [rubella] child is frequently intolerant of sound; recruitment is present in some cases, but is not invariable."[15]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(figure 1)[14]

Remediation

> In the 1964—65 rubella epidemic, between 20,000 and 40,000 babies whose mothers caught rubella in the early months of pregnancy were damaged to some degree. At least half of these children have impaired hearing, the most common and persistent defect found in rubella babies.[16]

A form of "remediation" is the abortion. When a woman knows that she had German measles early in her pregnancy, she may consider abortion. "Whether or not to perform an abortion in such cases is up to the attending physician and the parents. . . ."[17] Factors to be considered are, first of all, the mental health of the mother. Also the age of the fetus at the time the mother contracted rubella. Another point of consideration is the religious beliefs of the parents. The age of the parents and the ease of conception is also very important.[18]

> The control and treatment of viral infections center around three general approaches: Immunologic, chemotherapeutic, and host-resistant.[19]

It has been found that the most effective method has been specific immunization by means of vaccines. There have been several advances made in the research on vaccines to combat respiratory infections, mumps, rubella, and rubeola.

> Two strains of rubella vaccine are being produced commercially: the H. P. V. 77 strain of rubella vaccine developed at the National Institute of Health, and the Cendehill strain

> developed in Belguim. Trials with these vaccine strains have shown no side-effects when given to children. On the other hand possible risks of teratogenicity [the formation, or bringing forth, of monsters][20] when the vaccine is given to pregnant or about-to-become-pregnant women remains to be determined. Therefore, serologic tests to separate the 85 per cent to 90 per cent of women already immune need to standardize to select the 10 per cent to 15 per cent of childbearing-age women who should be immunized.[21]

If this were made a prerequisite for obtaining a marriage license, many congenital malformations could probably be prevented. It deserves equal emphasis with the universally required serology."[22]

> Meanwhile, the Department of Health, Education, and Welfare has an ambitious $16,299,000. program for immunization of children in 1969 to 1970, half of this amount being earmarked for rubella vaccination. Children being the chief carriers and disseminators of this terotogenic disease, it is hoped that rubella can be eliminated by a five year plan starting with children of one to two years of age, and then extending through the elementary school population.[23]

The rubella vaccination can also be given to adult women. A simple blood test is given first to see if immunization is necessary. If the person is not immune, then she may receive the vaccination. However, she is sternly warned that pregnancy is unadvisable for at least three months after the vaccine is given.

Since the rubella vaccine has been discovered, advertisements concerning it have come out. The advertisements tell the people what rubella is and what it can do to a developing fetus. (See figure 2 on page 8)[24]

Inoculate them Protect her

Rubella* means German measles. Although it is a minor ailment for children, rubella can be destructive if contracted by a pregnant woman. It can cause cerebral palsy and other birth defects. Now, children can be vaccinated for rubella. Through your support in the past, we were able to help in developing this new vaccine that will remove one of the causes of cerebral palsy. There are many more causes to be uncovered and conquered.

We still need your support.
Give to
United Cerebral Palsy.

*Every child aged one to eleven should have a rubella vaccination.

(figure 2)[24]

One television commercial shown in the New York area informed the people of the locations where free rubella vaccinations may be obtained.

"Deafness due to maternal rubella will, happily, become a rare curiosity within our lifetime."[25] "Prevention of this early hearing loss will bring us much closer to our ideal goal that every child is born with good hearing."[26]

III. RH FACTOR

It is estimated that about 15 per cent of the population have Rh negative blood type.[27] The other 85 per cent have Rh positive blood. Nothing abnormal happens in an Rh negative person unless his blood is mixed with Rh positive blood. This mixing of blood types usually occurs in two ways, blood transfusions or through the placenta in a pregnant woman.[28]

> . . . when an Rh negative mother has an Rh positive child, the mixture of these blood factors sometimes causes the mother to develop antibodies against the Rh positive blood of the child. Since it takes some time for these antibodies to develop, the first child is seldom affected by them. On the next pregnancy, however, it can happen that the antibodies may affect the child and cause damage to the blood cells. From this may result erythroblastosis, extreme jaundice, cerbral palsy or other abnormalities. We have found that a large percentage of the children with athetoid cerbral palsy have congenital hearing loss which results apparently from an Rh disparity.[29]

Goodhill also has stressed the importance of the Rh factor in the causation of congenital sensory-neural deafness.[30]

Affects on the Fetus

> Erythroblastosis fetalis is a disease of the newborn infant characterized by progressive agglutination and hemolysis of the red blood cells. agglutination—the clumping together of blood cells suspended in a fluid. hemolysis—the destruction of the red corpuscles with liberation of hemoglobin into the surrounding fluid.[31] In most instances of erythroblastosis fetalis the mother is Rh negative, the father is Rh posative, the baby has inherited the Rh positive characteristic from the father and the mother has developed anti-Rh agglutinins that have diffused into the baby to cause red blood cell agglutination.
>
> An Rh negative mother having her first Rh positive child usually does not develop sufficient anti-Rh agglutinins to cause any harm. However, an Rh negative mother having her second Rh positive child will have become 'sensitized' by the first child and therefore will often develop anti-Rh agglutinins very rapidly upon becoming pregnant with the second child.
>
> After anti-Rh antibodies have formed in the mother, they diffuse very slowly through the placental membrane into the baby's blood. There they cause slow agglutination of the baby's blood, and clumps of blood cells occlude small blood vessels. The red blood cells of these clumps gradually hemolyze, releasing hemoglobin into the blood. The reticuloendothelial system then converts the hemoglobin into bilirubin, which causes yellowness of the skin (jaundice). The antibodies also attack and damage many other cells of the body. The organ of corti included.[32]

Erythroblastosis fetalis (the Rh factor) causes anemia, deprivation of oxygen in the blood stream, and

destruction of cells and nerve tissue.[33] The fetus may develop cerebral jaundice and cerebral palsy.[34]

Remediation

> . . . erythroblastosis, which used to produce thousands of miscarriages, dead infants, and badly damaged babies, is now almost totally preventable.[35]
>
> The usual treatment for erythroblastosis fetalis is to replace the new-born infants blood with Rh negative blood. The Rh negative blood is infused while the Rh positive blood is removed. After the Rh negative blood is in the baby the destruction of red blood cells is over. By the time the Rh negative blood is replaced by the baby's own blood, the remaining anti-Rh agglutinins will have been destroyed.[36]

Some new research and experimentation has been successful in inter-uterin transfusions. These inter-uterin transfusions prevent the cell destruction which takes place at birth.

Blood transfusions into the tiny fetus in the mother's womb can be given on a regular basis. "A much less frequently used method is to operate on the uterus, free one of the fetal limbs and give a transfusion in that manner."[37]

The blood of the pregnant woman should be examined before her child is born. If the antibodies in her blood are high there is a great possibility that her child will be affected. "Prevention of the

sequelae of this type of blood dyscrasia is often possible."[38] Continual research on the Rh factor may someday make it a thing of the past.

IV. DRUGS

The senses of the body can be dulled in various ways by the use of different drugs or medicines. Also most of the functions of the body may be slowed because of some types of drugs. In the case of the senses the drugs usually operate through their affect on the nerves involved.

Of all the nerves of the body, the most sensitive are probably the auditory nerves. Some of the damaging drugs affect the nerves of the ear directly. Other drugs affect the hearing center of the brain. A fairly large number of rather common drugs possess a damaging property to the ear. It is quite likely that the over use of medication or the self inducement of these drugs may account for a large number of the hearing impairments in our country.[39]

> A number of drugs are known to be 'cochleotoxic', or damaging to the inner ear. In this category we can certainly place dihydroxtreplomycin. Kanomycin, Neomycin, and a few other injectable antibiotics that are particularly necessary for the treatment of serious bloodstream infections. Fortunately, the medical profession is becoming acutely aware of these dangers, and the utilization of these drugs is now limited to use in life-threatening conditions.[40]

It is not so well known that aspirin, quinine, and some other drugs may damage the inner ear and the hearing mechanism in some specifically sensitive people. It is also a possibility that nicotine and caffeine may affect the cochlear mechanism. More intensive study must be done on these commonly used drugs.[41]

Some researchers have shown ". . . experimentally that the ingestion of quinine produces hemorrhages in the inner ear of guinea pigs."[42] Others have tried to keep the use of quinine out of obstetrical practice, because it was thought that it caused congenital deafness. They stated that all suspected drugs should be completely avoided during pregnancy.[43]

Quinine was also used in childbirth, to induce labor. It was found that iodine was present in the brain of new born babies that were deaf. The mothers of these babies were administered quinine before their children were born.[44]

Proof has been found that the auditory nerve is more often affected by damaging substances in the bloodstream than any of the other nerves. Certain poisions seem to have a preference for the auditory nerve. Some people have a susceptability to these poisions.[45]

> The endolymph is secreted by epithelial cells in the stria vascularis. Streptomycin or quinine intoxication causes degeneration of the external

> and then internal hair cells followed by the disappearance of the whole organ of Corti and degeneration of the epithelial cells of the stria. This occurs in a few segments. The appearance of the mitochondria is altered in the hair cells and stria vascularis.[46]

Along with quinine, the salicylates, streptomycin and alcohol also pass through the placenta and may be found in the fetal circulation at the same strength as in the mothers bloodstream. They are therefore considered as a cause of deafness. Salicylates are widely used and are known to produce a lesion similar to that of quinine. Chronic alcoholism is also associated with congenital deafness.[47]

A late discovery has shown that thalidomide taken during early pregnancy results in severe congenital deformities often affecting the ear.[48]

> . . . there is no question that all suspected drugs should be completely avoided during pregnancy. Our ignorance of this entire subject of biologic deafness is so extreme that no safeguards against its development should be neglected whether or not actual supportive evidence is available.[49]

V. OTHER CAUSES

This brief history of prenatal causes of deafness should include some of the other, less common, influences on the embryo. Some of these causes are: emotional disturbances, accidents, influenza, diabetes, toxemia of pregnancy, and many others.

False Labor

"False labor is the circumstance of the mother behaving as though delivery is imminent but the birth does not materialize."[50] The birth process may not occur for a few days or weeks. These false alarms have occured with some frequency in the medical histories of children with hearing disorders.

> . . . The significance of false labor is obscure but several factors might be operative. Because of actual labor the placental blood supply might be disturbed during the false alarm causing deprivation. Toxic conditions also might be present at this time. False alarms might be indicative of undue anxiety in the mother with concomitant physiological implications. Moreover, undue anxiety might cause increased tensions when the birth does occur.[51]

Influenza

"Other viral infections, notably influenza, are thought to account for a small number of cases."[52] If the influenza occurs during the first three months of pregnancy, it may cause deafness in the developing embryo. "Asian influenza has also been noted as a cause of deafness. . . ."[53]

"The child deaf from maternal influenza bears some resemblance to the child deaf from maternal rubella; both are small, and a little slow in development."[54]

Congenital Syphilis

Congenital syphilis is included in the prenatal group of causes of deafness even though the symptoms of deafness are rarely present at birth. ". . . the noxious influences of the syphilitic disease are transmitted from the infected mother to her child during her pregnancy."[55] Sometimes the deafness begins in the first two years of life. This is an early form. In a late form the deafness occurs between the ages of eight and twenty. Often it occurs at the time of puberty.[56]

Diabetes

Having diabetes during pregnancy is recorded as an occasional cause of deafness. "It is surprising that it is not more common in view of the tendency of diabetics to develop peripheral neuritis."[57]

Beri-Beri

Beri-beri is a disease caused by a vitamin B_1 deficiency in the diet. Its characteristics are extreme weakness, paralysis, and anemia.[58] Hearing loss has been found in babies born of women suffering from beri-beri during pregnancy.[59]

Glandular Fever

Bilateral deafness has been found in children born of women suffering from glandular fever during pregnancy.[60]

Cretinism

Cretinism is a condition of the thyroid. "This condition of hypothyroidism begins during fetal life and may be endemic or sporadic."[61]

The characteristics of this syndrome are dwarfism, mental deficiency and backward in all development. Skin abnormalities and deafness are also characteristics of Cretinism.[62]

Malabsorption Syndrome

The malabsorption syndrome is suspected as a cause of sporadic congenital deafness. It is present when their is a lack of vitamin A in the diet of the mother during her pregnancy. This lack of vitamin A causes many defects in the development of the fetus.[63]

Toxemia of Pregnancy

About seven per cent of all pregnant women have very rapid weight gain during pregnancy. This is known as toxemia of pregnancy, and it is caused by an over retention of salt by the kidneys.[64]

"The association of toxema of pregnancy with some causes of congenital deafness is now clinically accepted. The signs of the toxema in some cases have been very mild."[65]

IV. CONCLUSION

"Congenital deafness may be caused by almost any severe virus infection of the expectant mother during the early months of pregnancy."[66] It may also be caused by such things as: the Rh factor, certain drugs, many miscellaneous accidents in pregnancy, and various diseases in early pregnancy. The first three months of pregnancy are the most important time in the development of the fetus. During this period the eyes, ears, and central nervous system are rapidly being formed. Any injury or accident at this time may be very critical.[67]

> Interesting and exciting possibilities are constantly presenting themselves to those engaged in research, and it is the earnest hope of all who work for the deaf child that more and more cases of congenital deafness will soon be preventable.[68]

FOOTNOTES

[1]John Chalmers Ballantyne, Deafness (Boston: Little, Brown, and Company, 1960), p. 124.

[2]Rubolf Pint-Jon Eisenson and Mildred Slanton, Hearing and Deafness (Appleton-Centure-Crofts, Inc., 1941), p. 103.

[3]Edith Whetnall and D. B. Fry, The Deaf Child (Springfield, Ill.: Charles C. Thomas, 1964), p. 95.

[4]Edna Simon Levine, The Psychology of Deafness (New York: Columbia University Press, 1960), p. 315.

[5]"Measles Threat to Unborn," Science News Letter, September 26, 1964, p. 194.

[6]Norton Canfield, Hearing, A Handbook for Laymen (New York: Doubleday and Company Inc., 1959), p. 57.

[7]Ibid.

[8]Whetnall, p. 111.

[9]Ballantyne, p. 131.

[10]Ibid., p. 109

[11]Ibid., p. 128.

[12]Ibid., p. 109.

[13]Ibid., p. 131.

[14]Ibid., p. 157.

[15]Whetnall, p. 111.

[16]Richard L. Masland, "Rubella Can Rob Children of Their Hearing," Volta Review, LXX (May, 1968), 304.

[17]Science News Letter, p. 194.

[18]Ibid.

[19]Wallace E. Herrell, "Progress in Control of Viral Infections," Volta Review, LXX (December, 1968), p. 95.

[20]Hoerr Jones, Blakinston's New Gould Medical Dictionary (Philadelphia: The Blakiston Company, 1949), p. 1033.

[21]John H. Talbott and Robert N. Mayo, "Rubella Vaccination," Archives of Otolaryngology, XC (July, 1969), p. 3.

[22]Herrell, p. 707.

[23]Talbott, p. 3.

[24]Glamour, LXII (February, 1970), p. 68.

[25]Talbott, p. 3.

[26]Canfield, p. 57.

[27]Helmer R. Myklebust, The Psychology of Deafness (New York: Grune and Stratton, 1954), p. 36.

[28]Joseph Sataloff, "The Rh Factor in Congenital Deafness," Volta Review, LV (July, 1969), p. 311.

[29]Ibid.

[30]Myklebust, The Psychology of Deafness, p. 37.

[31]David B. Guralnik, Webster's New World Dictionary of the American Language (Nashville: The Southwestern Company, 1966), pp. 14 and 349.

[32]Arthur C. Guyton, Textbook of Medical Physiology (Philadelphia: W. B. Saunders Company, 1961), p. 193.

[33]Myklebust, The Psychology of Deafness, p. 37.

[34]Joseph Sataloff, Hearing Loss (Philadelphia: J. B. Lippincott Company, 1966), p. 379.

[35]Ellen Switzer, "Childbirth Advances—more in the last five years than in the previous hundred," Glamour, LXII (February, 1970), p. 217.

[36]Guyton, p. 194.

[37]Switzer, p. 218.

[38]Boris V. Markovin, Through the Barriers of Deafness and Isolation (New York: The Macmillan Company, 1960), p. 15.

[39]A. F. Niemoeller, Complete Guide for the Deafened (New York: Harvest House, 1960), p. 65.

[40]Victor Goodhill, "Deafness Research: Where are we?", Volta Review, LXX (November, 1968), p. 627.

[41]Ibid.

[42]Walter Hughson and Eva Thompson, "Otological Research and Its Recent Contributions to Clinical Otology," Annals of Otology, Rhinology and Laryngology, L (March, 1941), p. 95.

[43]Ibid.

[44]Niemoeller, p. 66.

[45]Whetnall, p. 113.

[46]Ibid.

[47]Ibid.

[48]Ibid.

[49]Ibid.

[50]Helmer R. Myklebust, Auditory Disorders in Children (New York: Grune and Stratton, 1954), p. 57.

[51]Ibid., p. 58.

[52]Ballantyne, p. 128.

[53]Whetnall, p. 112.

[54]Ibid.

[55]Ballantyne, p. 129.

[56]Ibid.

[57]Whetnall, p. 112.

[58]Guralnik, p. 70.

[59]Whetnall, p. 112.

[60]Ibid.

[61]Ibid., p. 113.

[62]Ibid.

[63]Ibid., p. 112.

[64]Guyton, p. 1007.

[65]Whetnall, p. 113.

[66]Hallowell Davis and Richard S. Silverman, Hearing and Deafness (New York: Holt, Rinehart, and Winston, Inc., 1962), p. 109.

[67]Ibid.

[68]Ballantyne, p. 131.

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